Cystic Fibrosis in Australia 2014

17TH ANNUAL REPORT AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY

CYSTIC FIBROSIS Australia

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17th Annual Report from the Australian Cystic Fibrosis Data Registry

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Preface

I am very pleased to deliver this 17th annual report from the Australian Cystic Fibrosis Data Registry, for the year 2014.

For the first time we have crossed an important demographic threshold: more than half of the cystic fibrosis (CF) population in Australia is adult. So much progress has been made in the last decade and a half in the care and treatment of people with CF. Only one third of Australians with CF were adult in 1998 when the Registry began. Last year the proportion stood tantalisingly close at 49.9 per cent but this report's finding that this has grown to 51.1 per cent.

Not all centres have reported details of the social status of their adult patients. In those that have, two thirds of patients were in full time or part time work; almost 4 out of 10 had post school qualifications and a similar proportion were married or in de facto relationships. One in eight males and almost one in five females have at least one child.

With increasing access to new therapies, the opportunity for more Australians with CF to have normal adult lives is becoming real. G551D mutation are the second most common mutations, carried by 203 people aged 6 years and over. These people are eligible for treatment with Kalydeco, a drug that has had a remarkable impact on the lives of people with CF. Orkambi, which combines Kalydeco with another agent (lumacaftor) has the potential to improve the lives of those with two copies of the F508del mutation. It is currently available to CF populations in North America and parts of Europe for people aged over 12 years with two copies of the F508del mutation. If approval was mirrored in Australia, as is Cystic Fibrosis Australia's (CFA) profound wish, a further 1,024 people might benefit. Future annual reports from the Registry will map the progress that is expected to result from introducing these drugs.

I thank the CF teams across the country who not only devote themselves to providing excellent care to their patients, but also for their time in contributing the data. Their efforts record progress and, through reporting and analysis, contribute to the cycle of improvement in the CF care and therefore in CF lives.

Nettie Burke
Chief Executive Officer
Cystic Fibrosis Australia
June 2016

Acknowledgements

Many thanks go to the ACFDR Advisory Committee, whose members are:

Dr Scott Bell – The Prince Charles Hospital, Brisbane QLD
Dr Peter Bye – Royal Prince Alfred Hospital, Camperdown NSW
Dr Peter Cooper – The Children's Hospital, Westmead NSW
Dr Adam Jaffe – Sydney Children's Hospital, Randwick NSW
Dr Guy Marks – Liverpool Hospital NSW
Dr James Martin – Women's and Children's Hospital, Adelaide SA
Dr Sarath Ranganathan – Royal Children's Hospital, Melbourne VIC
Dr Phil Robinson – Royal Children's Hospital, Melbourne VIC
Dr Gerard Ryan – Sir Charles Gairdner Hospital, Perth WA

Further acknowledgement must go to the following people and organisations:

Ms Ann-Maree Bosch for logistical arrangements and minutes of proceedings of the Advisory Committee

Mr Geoff Sims of Australian Clinical Registries for database management and reporting

CFA also acknowledges generous support from Vertex, Mylan, Roche, Novartis and the participants from the Cystic Fibrosis Australia Great Escape

Participating Centres

The ACFDR relies on the tireless work of people in the following CF Centres who enter data and handle edit queries for quality control of the annual collection of data:

New South Wales
Sydney Children's Hospital, Randwick
Royal Prince Alfred Hospital, Sydney
The Children's Hospital, Westmead
Westmead Hospital – Adults
Gosford Hospital, Gosford
John Hunter Children's Hospital, Newcastle
John Hunter Hospital - Adults, Newcastle

Victoria
Royal Children's Hospital, Melbourne
The Alfred Hospital, Melbourne
Monash Medical Centre, Clayton

Queensland
Royal Children's Hospital, Brisbane
The Prince Charles Hospital, Brisbane
Mater Hospital – Children, Brisbane
Mater Hospital – Adults, Brisbane
Gold Coast University Hospital – Southport

South Australia
Royal Adelaide Hospital, Adelaide
Women's and Children's Hospital, Adelaide

Western Australia
Princess Margaret Hospital for Children, Perth
Sir Charles Gairdner Hospital, Perth

Tasmania
Royal Hobart Hospital, Hobart
Launceston General Hospital, Launceston
North West Regional Hospital, Burnie

Australian Capital Territory
The Canberra Hospital, Garron

Contents

1 People with cystic fibrosis

1.1 Overview

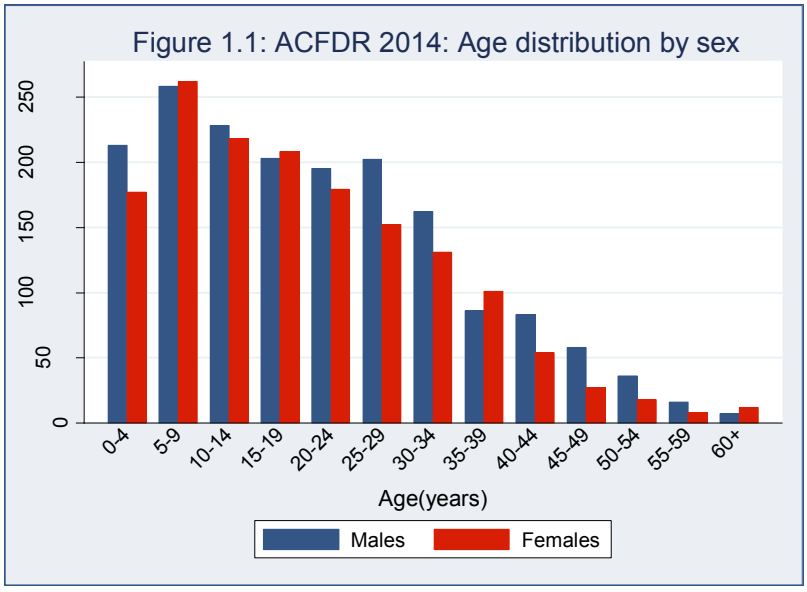
At 31 December 2014 the Australian Cystic Fibrosis Data Registry (ACFDR) held records of 3,294 people with cystic fibrosis, 59 more than at the end of 2013.

The mean age of the registry population was 20.5 years, increased from 20.0 years reported in 2013. Reflecting a steady upward trend in age of Australians with CF, the proportion of the Registry population that is adult (18 years and over) increased to 51.1 per cent in 2014, from 49.9 per cent in 2013. For the first time, more than half of the Australian CF population as recorded by the Registry is adult. Only one third of patients were adult in 1998, when all major centres first contributed data to the Registry.

The median age of 18.4 years at 31 December 2014 is also greater than at the end of previous years, having been 17.9 in 2013 and 17.7 in 2012. Median age for males (19.2 years) remained higher than that for females (17.7 years) in 2014.

An increase of 59 in the overall number of registrants in 2014 is approximately equal to the excess of new diagnoses (79) over deaths (19) reported for the year.

1.2 Age distribution



ACFDR 2014: Age and sex of registrants at 31 December 2014

Age group	Males	Females	Persons	Per cent male
Standard demographic age groups:				
0 - 4 years	213	177	390	54.6
5 - 9 years	258	262	520	49.6
10 - 14 years	228	218	446	51.1
15 - 19 years	203	208	411	49.4
20 - 24 years	195	179	374	52.1
25 - 29 years	202	152	354	57.1
30 - 34 years	162	131	293	55.3
35 - 39 years	86	101	187	46.0
40 - 44 years	83	54	137	60.6
45 - 49 years	58	27	85	68.2
50 - 54 years	36	18	54	66.7
55 - 59 years	16	8	24	66.7
60 + years	7	12	19	36.8
Alternative CF age groups and totals:				
0 - 1 years	86	65	151	57.0
2 - 5 years	165	161	326	50.6
6 - 11 years	291	307	598	48.7
12 - 17 years	284	251	535	53.1
Children and adolescents	*826*	*784*	*1610*	*51.3*
18 - 29 years	473	412	885	53.4
30 + years	448	351	799	56.1
Adults	*921*	*763*	*1684*	*54.7*
Total, all ages	*1,747*	*1,547*	*3,294*	*53.0*

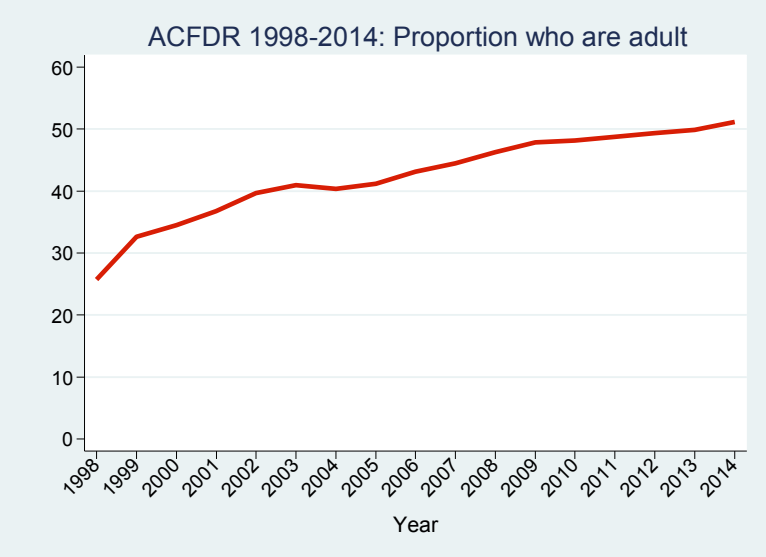
The lower table area shows age categories recommended for international comparison of CF data. Many of the tables and charts later in this report use these age categories.

At 31 December 2014, males made up 53.0 per cent and females 47.0 per cent of the ACFDR population. This has remained a consistent proportion since establishment of the Registry in 1998. The proportion of males is higher among the adult population (54.7%) than the child and adolescent population (51.3%). In line with international experience, there is better survival of males in the Australian CF population.

The proportion of adults in the Registry as a whole was 51.1 per cent at 31 December 2014. The proportions for States and Territories are shown in the following table, although those for smaller jurisdictions should be interpreted in the context of their smaller populations.

ACFDR 31 December 2014: Adult status by State/Territory of residence

State or Territory of residence	Child/adolescent	Adult	Total	Per cent adult
New South Wales	471	502	973	51.6
Victoria	313	358	671	53.4
Queensland	414	424	838	50.6
Western Australia	193	176	369	47.7
South Australia	145	141	286	49.3
Tasmania	49	54	103	52.4
Australian Capital Territory	20	22	42	52.4
Northern Territory	3	5	8	62.5
Overseas	2	2	4	50.0
Total	*1,610*	*1,684*	*3,294*	*51.1*



1.3 Family, adult marital status, education and activity

The following needs to be interpreted in the light of under-reporting of social characteristics by some CF centres, as noted for each set of data below.

Data from CF centres reporting sibling status for at least two thirds of their patients showed that 17 per cent of paediatric patients and 22 per cent of adult patients were known to have siblings with CF. These proportions are calculated from around 55 per cent of the total CF population.

Thirty seven per cent of male adult patients and 41 per cent of adult female patients for whom marital status was reported were married or in defacto relationships. Three Centres that did not report marital status for more than one-third of their patients are excluded from this analysis.

Twelve per cent of adult male CF patients and 18 per cent of adult females had at least one child. These calculations exclude patients at two large adult CF centres and three medium or smaller centres that make up around 43 per cent of the adult CF population.

ACFDR 31 December 2014: Marital status of adults[a]

	Males		Females	
Marital status	**Number**	**Per cent**	**Number**	**Per cent**
Married (includes de facto)	215	36.7	194	40.8
Not married	371	63.3	281	59.2
		100.0		*100.0*
Unknown or missing	97	14.2	*93*	16.4
Total	*683*		*568*	

(a) Centres with missing data for more than one third of adult patients were excluded. They comprised one large, one medium size and one smaller adult CF centre.

Many people with cystic fibrosis continue with education beyond senior secondary school level. Just over 15 per cent of adult CF patients for whom educational attainment was reported having university qualifications and a further 23 per cent having completed other study beyond high school. Data presented in the table exclude centres that did not report education attainment for more than half of their patients, representing about half of the adult patients in the Registry.

ACFDR 31 December 2014: Educational attainment of adults[a]

	Number	**Per cent**
Junior Secondary (Year 10)	88	14.0
Senior Secondary (Year 12)	298	47.3
Tertiary certificate or diploma	145	23.0
University degree	95	15.1
Left school prior to Year 10	4	0.6
Total reported	*630*	*100.0*
Unknown/not reported (incl. as % of total below)	195	23.6
Total	*825*	*100.0*

(a) CF centres with educational attainment reported for less than 50 per cent of patients were excluded from analysis. They comprised two large adult CF centres and six medium or small centres.

Sixty seven per cent of adults with CF for whom activity status was reported were in either full time or part time paid employment during 2014. Around 43 per cent of the adult CF population was excluded from this analysis.

ACFDR 31 December 2014: Activity status of adults [a]

	Number	**Per cent**
Employed, full time paid	284	36.1
Employed, part time paid	241	30.7
Voluntary work only	4	0.5
Unemployed	39	5.0
Pensioner	46	5.9
Others not in labour force (b)	172	21.9
Total reported	*786*	*100.0*
Unknown/not reported (incl. as % of total below)	171	17.9
Total	*957*	*100.0*

(a) CF centres with missing activity status data for more than one third of patients were excluded from analysis. They comprised two large adult CF centres and four medium or small centres.
(b) includes homemakers, students

2 Diagnosis

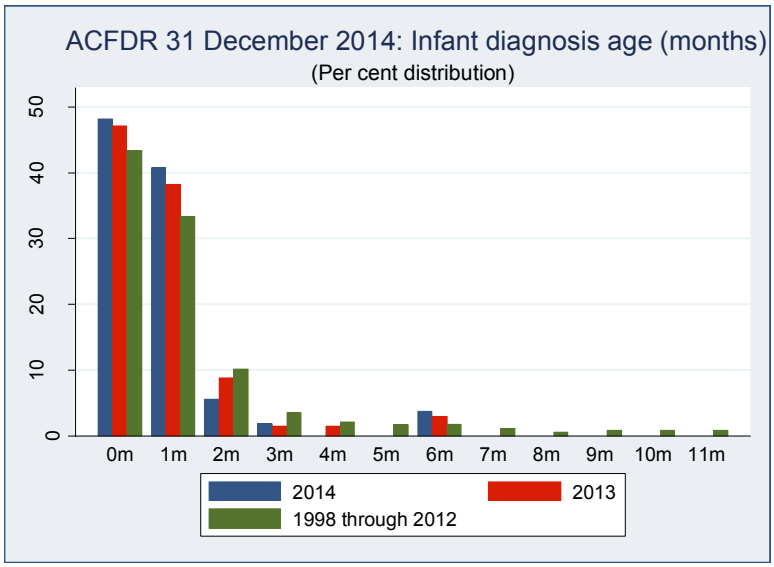
2.1 Overview

There were 79 new diagnoses of cystic fibrosis (CF) notified to the Registry for 2014, including 64 patients who were diagnosed at less than one year of age.

Infant diagnoses are not always made or reported in the year of birth and numbers vary across years. An estimate of CF birth incidence can be made using birth cohorts from earlier birth years, say 2009 to 2013, to avoid the reporting lag and to allow for annual variation. Using the average number aged from 1 to 5 years in 2014 (84) and the average number of births reported by the Australian Bureau of Statistics for their birth years (305,000 across years 2009 to 2013), an estimate of Australian birth incidence for CF is 28 per 100,000 or 1 in 3,630 births.

2.2 Age at diagnosis

All but 3 of the infant diagnoses where a diagnosis date was reported (54 out of the 64 new infant diagnoses) were completed by three months of age, assisted by neonatal screening programs that operate in all States and Territories of Australia. There were 10 infant diagnoses in 2014 where a diagnosis date was not reported but where the fact of infant diagnosis was inferred from their age being less than 12 months at the end of the year.



Australian CF centres reported 6 new cases diagnosed in early childhood (1 to 4 years), 3 aged from 5 to 9 years, 2 in the age group 14 to 24 years and 4 at ages 35 years and over.

2.3 Presentation and diagnosis

Over 60 per cent of new cases of CF diagnosed in 2014 included neonatal screening as a mode of presentation and 13 per cent reported meconium ileus. Respiratory symptoms were reported in 10 per cent, and gastrointestinal symptoms in 1.3 per cent.

ACFDR 31 December 2014: Mode of presentation [(a)] by year of diagnosis

	All years	2014	All years	2014
	Number		*Per cent*	
Neonatal screening	1,687	48	53.8	60.8
Respiratory symptoms	462	8	14.7	10.1
Gastrointestinal symptoms	358	1	11.4	1.3
Meconium ileus	417	11	13.3	13.9
CF sibling	254	5	8.1	6.3
Minor manifestations	29	0	0.9	0.0
Pre-natal diagnosis	46	1	1.5	1.3
Infertility	18	0	0.6	0.0
Other	325	7	10.4	8.9
Unknown	160	0	0.0	0.0
Total	*3,294*	*79*	*100.0*	*100.0*

(a) More than one mode of presentation can be recorded for a patient so numbers in this section add to more than the total number of registrants and percentage columns add to more than 100.0.

2.4 Phenotype

The proportion of patients who are pancreatic insufficient is 82.9 percent, based on consolidated data across all years of reporting and excluding just under 5 per cent of patients for whom pancreatic status is unknown or is missing.

Sweat chloride values have been reported for just over half (54%) of patients in the Registry. Of these, there were 199 patients for whom sweat chloride values were below 60 mmol/L, three out of 10 of whom had at least one copy of the R117H mutation. Of the 34 patients whose sweat chloride values were below 30 mmol/L, 7 out of 10 had a copy of the R117H mutation.

2.5 Genotype

Mutation information consolidated across reporting years was available for 3,035 patients, or 92.1 per cent of all patients in the Registry at the end of 2014.

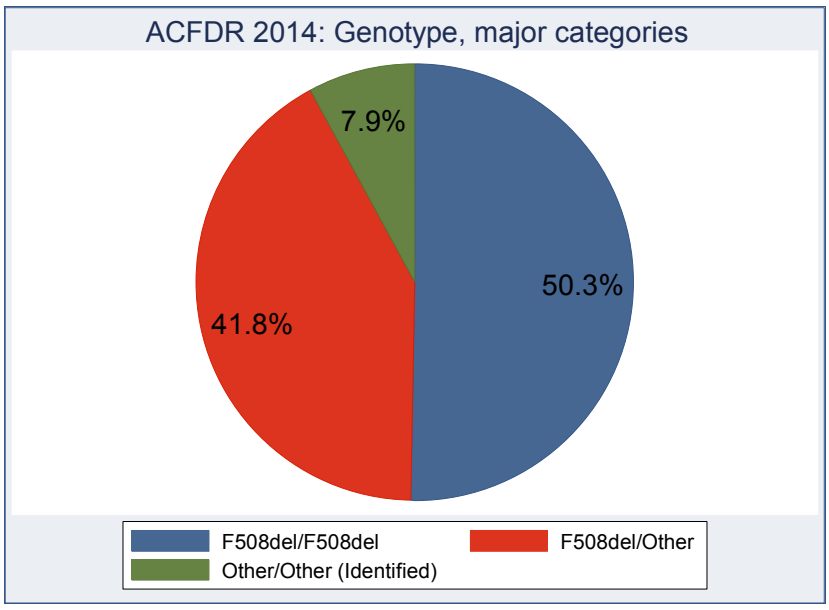
The genetic mutation F508del has been identified as at least one of the paired mutations responsible for the inheritance of cystic fibrosis in 92.1 per cent of patients for whom genotype details have been reported. Over half of the total, 1,526 patients (50.3%) are reported as homozygous for F508del, with 1,024 of these being aged 12 years and over.

G551D was the next most prevalent mutation, with 231 or 7.6 per cent of the CF population having this mutation, mostly in combination with F508del or another mutation. Of these patients, 203 were aged 6 years and over.

ACFDR 31 December 2014: Genotype[(a)]

	Mutation 1										
	F508 del	G 542 X	G 551 D	N 1303 K	W 1282 X	R 117 H	1717 -1G ->A	621 +1G ->T	Other NEC	Unk-nown	Total
	Per cent										
Mutation 2:											
F508del	50.3										50.3
G542X	1.9	0.1									2.0
G551D	6.0	0.2	0.2								6.4
N1303K	1.1	0.1	0.1	0.1							1.3
W1282X	0.6	0.0	0.0	0.0	0.2						0.8
R117H	3.3	0.0	0.1	0.0	0.0	0.1					3.5
1717-1G->A	1.3	0.0	0.0	0.0	0.0	0.0	0.0				1.4
621+1G->T	1.2	0.1	0.0	0.0	0.0	0.0	0.0	0.0			1.3
Other NEC	15.4	0.3	0.5	0.1	0.1	0.2	0.2	0.2	1.6		18.6
Unknown	11.1	0.3	0.4	0.0	0.0	0.1	0.0	0.0	1.1	1.4	14.5
Total	*92.1*	*1.0*	*1.4*	*0.2*	*0.3*	*0.5*	*0.2*	*0.3*	*2.7*	*1.4*	*100.0*

(a) Patients with missing genotype data for both alleles were excluded from analysis



The table over the page shows population and allele prevalence of the most common cystic fibrosis transmembrane conductance regulator (CFTR) mutations found in the Australian CF population. A more extended list is available on request. Just 21 mutations have a population prevalence of 10 or more.

ACFDR 2014: Patients and alleles – most common CFTR mutations in Australian CF population

CFTR Mutation	Patient Number	Patient Per cent	Homozygous Patient Number	Allele Number
F508del	2,795	92.1	1,526	4,321
G551D	231	7.6	7	238
R117H	119	3.9	3	122
G542X	88	2.9	3	91
1717-1G->A	47	1.5	0	47
621+1G->T	46	1.5	1	47
N1303K	43	1.4	2	45
W1282X	27	0.9	5	32
R553X	23	0.8	1	24
5T	22	0.7	0	22
D1152H	18	0.6	0	18
P67L	17	0.6	0	17
2789+2insA	15	0.5	0	15
Q493X	14	0.5	0	14
E60X	11	0.4	1	12
V520F	11	0.4	0	11
R1162X	10	0.3	0	10
G85E	10	0.3	0	10
R560T	10	0.3	0	10
3272-26A->G	10	0.3	0	10
R334W	10	0.3	0	10
1078delT	10	0.3	0	10
2789+5G->A	9	0.3	0	9
I507del	9	0.3	0	9
1898+1G->A	9	0.3	0	9
3659delC	9	0.3	0	9
A455E	9	0.3	0	9
3849+10kbC->T	8	0.3	0	8
2183AA->G	5	0.2	3	8
R347P	8	0.3	0	8
2184delA	8	0.3	0	8
1154insTC	8	0.3	0	8
S549N	8	0.3	0	8
R1066C	6	0.2	0	6
R75Q	4	0.1	1	5
711+3A->G	5	0.2	0	5
3121-1G->A	4	0.1	1	5
394delTT	4	0.1	0	4
R347H	4	0.1	0	4
R117C	4	0.1	0	4
R352Q	4	0.1	0	4
I1027T	4	0.1	0	4
2622+1G->A	4	0.1	0	4
3791delC	4	0.1	0	4
R1070W	4	0.1	0	4
Other mutations , not listed above	292	9.6		
Unknown mutation	440	14.5		
Total patients genotyped	*3,035*	*100.0*		

3 Health and functioning

Information in this chapter covers respiratory infections, medical complications, lung function and nutritional measures. Two adult centres did not submit microbiology information for their patients in 2014 and are excluded from the analysis. All centres submitted data for multiple occasions where clinical measures of height, weight and lung function were taken.

3.1 Respiratory infections

Patients who were tested for respiratory infections in 2014 had a mean of 3.7 tests of all types during the year. The median number of tests was 3 overall, and 4 in the age groups between 6 and 17 years. Two adult centres were excluded from this analysis.

ACFDR 2014: Number of sputum and BAL/bronchoscopy cultures[a]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
			Per cent of patients tested (b)				
Sputum cultures:							
None	56.0	31.9	14.4	5.2	1.2	0.9	11.0
1	7.0	13.6	15.3	16.2	26.6	30.4	20.4
2	9.0	8.9	11.6	10.0	22.0	18.3	14.7
3	7.0	8.1	9.5	11.3	12.0	14.1	11.0
4	5.0	10.6	11.4	11.5	8.7	9.6	10.0
5	2.0	6.8	10.4	9.4	8.1	7.6	8.3
6	6.0	8.9	7.4	11.0	6.4	3.9	7.3
7 or more	8.0	11.1	20.0	25.4	15.1	15.2	17.3
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
BAL/bronchoscopy:							
None	56.0	63.8	88.4	96.9	98.7	97.8	89.8
1	34.0	29.8	10.2	2.9	0.6	1.4	8.3
2	8.0	6.0	1.4	0.3	0.6	0.9	1.7
3 or more	2.0	0.4	0.0	0.0	0.2	0.0	0.2
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
			Number of patients				
Patients tested (b)	*100*	*235*	*431*	*382*	*518*	*355*	*2,021*
Culture not done	51	91	167	153	169	243	874
Total reported	*120*	*274*	*526*	*472*	*660*	*544*	*2,596*
Not reported	31	52	72	63	27	54	299
Total patients	*151*	*326*	*598*	*535*	*687*	*598*	*2,895*

(a) Two adult CF centres were excluded from analysis
(b) By any method of obtaining culture.

The table on the previous page shows the distribution of CF patients according to the number of both sputum and BAL/bronchoscopy samples examined during 2014. The latter method is used mainly on smaller children. Taking sputum samples alone, more than two thirds (69 per cent) of the patients tested had at least two sputum samples in 2014. Respiratory cultures were not performed for one third of patients with either test results or 'not tested' reported. It can also be seen that respiratory culture information was not reported at all for around one in ten patients.

The most commonly identified organisms in respiratory specimens are various species and forms of *Pseudomonas*. It can be seen that 48.5 per cent of patients tested produced positive *Pseudomonas aeruginosa* cultures, with the mucoid form showing in 32.0 per cent. Its prevalence is greater in adult patients, with 60.9 per cent of tested adult CF patients producing samples indicating the mucoid form of *Pseudomonas aeruginosa*, three times the corresponding proportion for adolescents and much higher than that for children.

ACFDR 2014: Pseudomonas infection by age group[(a)(b)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Pseudomonas aeruginosa:							
Mucoid	1.0	1.3	8.1	20.2	53.9	71.0	32.0
Rough/non-mucoid	7.0	7.7	16.7	24.9	33.6	33.5	24.0
Not differentiated	6.0	6.0	11.8	12.8	18.3	22.5	14.6
Any Ps aeruginosa	14.0	14.0	28.5	40.3	70.3	82.3	48.5
Pseudomonas other species	0.0	3.0	2.3	2.6	2.5	1.1	2.2
				Number of patients			
Patients tested	*100*	*235*	*431*	*382*	*518*	*355*	*2,021*

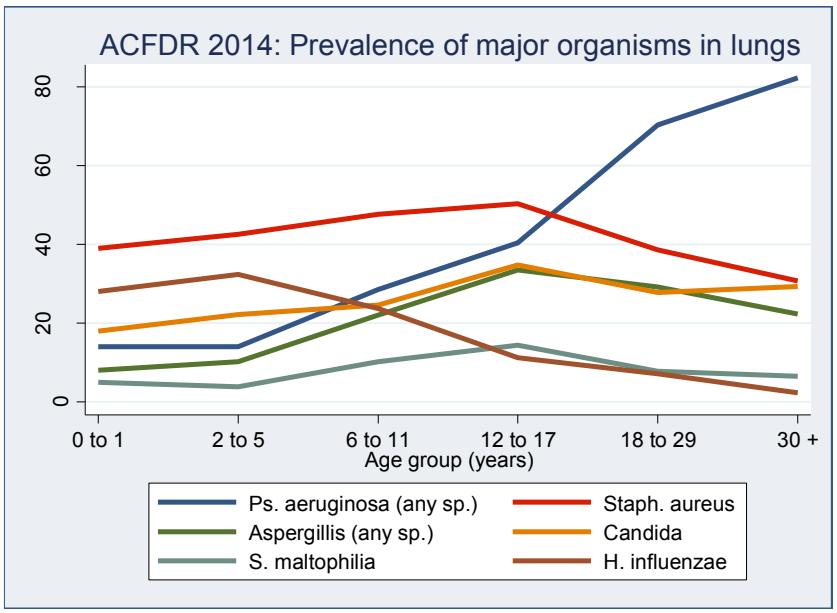
(a) Two adult CF centres were excluded from analysis
(b) Patients may have had more than one type of Pseudomonas infection. Percentages for individual types may add to more than totals.

While prevalence of Pseudomonas organisms is lower in children than in adults, (see table above), young children are more likely than adult patients to produce cultures showing presence of Staphylococcus aureus (see table, next page). Half of all children and adolescents aged 6 to 17 years had this bacterial infection. Haemophilus influenzae is also evident in relatively high proportions of children, especially in children aged from 2 to 5 years, where this organism was cultured for almost one third of children. The youngest age groups also had the highest proportions with positive cultures of the bacteria Escherichia coli; 12 per cent, for those in the age group less than 2 years, being the highest.

ACFDR 2014: Other respiratory culture by age group(a)

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
			Per cent of patients tested (b)				
Bacteria:							
Staphylococcus aureus	39.0	42.6	47.6	50.3	38.6	30.7	41.8
Haemophilus influenzae	28.0	32.3	23.7	11.3	7.1	2.3	14.5
Burkholderia cepacia (Ps cepacia)	0.0	0.0	1.6	2.1	4.2	2.8	2.3
Stenotrophomonas maltophilia	5.0	3.8	10.2	14.4	7.7	6.5	8.7
Escherichia coli	12.0	8.9	6.5	3.1	1.0	0.6	4.0
MRSA (c)	2.0	1.7	1.9	3.4	2.5	3.4	2.6
Alcaligenes xylosoxidans	0.0	0.4	3.0	2.6	5.0	5.6	3.5
Serratia marcescens	2.0	1.7	0.9	1.3	0.8	0.0	0.9
Klebsiella (any species)	9.0	3.0	0.7	1.6	0.6	1.1	1.6
Non-tuberculous mycobacterium	0.0	0.0	0.0	3.1	4.6	4.2	2.5
Fungi:							
Candida	18.0	22.1	24.6	34.8	27.8	29.3	27.6
Aspergillus (any species)	8.0	10.2	22.0	33.5	29.2	22.3	24.0
Scediosporium (any species)	0.0	0.4	2.6	6.3	5.0	3.4	3.7
Other organisms not listed above	28.0	33.2	30.4	29.1	20.8	19.7	26.0
Normal flora only	63.0	79.1	82.8	77.5	38.0	28.5	59.4
No growth/sterile culture	*10.0*	*8.1*	*7.0*	*4.5*	*4.2*	*3.4*	*5.4*
			Number of patients				
Patients tested	*100*	*235*	*431*	*382*	*518*	*355*	*2,021*

(a) Two CF Centres were excluded from analysis
(b) Note: Patients may have multiple infections during the year. Percentages may add to more than 100.0.
(c) Methicillin-resistant Staphylococcus aureus



3.2 Other medical complications

Prevalence of medical complications increases with age in CF patients. For instance, 44 per cent of adult patients suffer gastro-oesophageal reflux, over one quarter of patients aged 30 years and over experience chronic insulin-dependent diabetes and over 40 per cent of the same age group have osteoporosis or osteopenia.

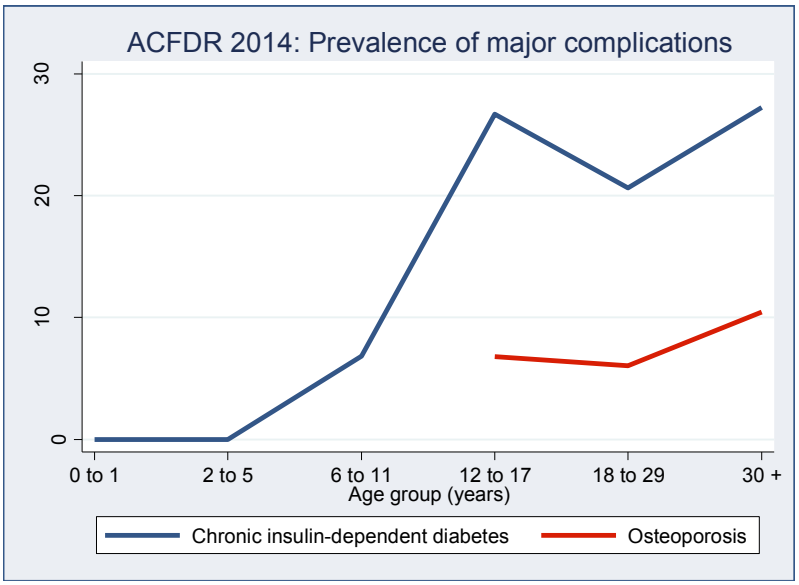
The proportion for whom none of the selected complications shown in the following table have been reported is over 88 per cent for children under 6 years, but declines to 16 per cent in CF patients aged 30 and over.

ACFDR 2014: Medical complications[(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
				Per cent			
Pulmonary:							
Major haemoptysis	0.0	0.7	0.4	0.8	4.8	8.7	3.7
Massive haemoptysis	0.0	0.0	0.0	0.0	0.2	1.7	0.5
Therapeutic bronchial artery embolisation	0.0	0.0	0.0	0.4	1.0	2.3	0.9
Pneumothorax	0.0	0.0	0.0	0.4	1.9	2.0	1.1
Any pulmonary above	0.0	0.7	0.4	1.6	6.9	12.8	5.4
Gastro-intestinal:							
Gastro-oesophageal reflux	4.6	4.3	10.4	16.7	41.5	47.5	28.8
- proven at endoscopy	0.0	0.7	1.2	3.6	5.0	5.8	3.7
Abnormal liver function test	6.2	6.5	11.2	15.1	25.6	26.1	19.1
Cirrhosis or portal hypertension	0.0	0.7	2.4	7.2	4.2	2.3	3.5
Pancreatitis	0.0	0.7	0.8	0.4	2.3	2.6	1.6
Any Gastro-intestinal above	10.8	10.1	19.7	30.3	59.0	61.2	41.9
Endocrine:							
Chronic insulin-dependent diabetes	0.0	0.0	6.8	26.7	20.6	27.2	18.1
Intermittent insulin-dependent diabetes	0.0	0.0	0.8	0.8	4.0	2.9	2.2
Other glucose abnormality	1.5	0.0	7.2	17.5	19.4	15.7	13.7
Any Endocrine above	1.5	0.0	13.3	40.6	41.5	44.3	31.9
Osteo:							
Osteoporosis	0.0	0.0	0.0	6.8	6.0	10.4	5.4
Osteopenia	0.0	0.0	1.6	15.1	22.1	31.0	16.7
Fracture this year	0.0	0.0	0.0	0.4	1.3	2.0	0.9
Any Osteo above	0.0	0.0	1.6	15.1	27.1	40.6	20.4
Other:							
Cancer	0.0	0.0	0.4	0.4	0.6	1.7	0.7
None of the above	87.7	89.2	69.5	38.6	20.2	15.7	39.4
Total reported (b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total reported	*65*	*139*	*249*	*251*	*480*	*345*	*1,529*
Unknown or not stated	60	169	337	279	186	179	1210
Total patients (a)	*125*	*308*	*586*	*530*	*666*	*524*	*2,739*

(a) Two adult CF centres were excluded from analysis
(b) Patient may have had more than one complication. Percentages add to more than 100.0.

Although some prevalence of osteoporosis at younger ages is reported in the table above, this is not displayed on the following chart because of uncertainty about diagnosis at younger ages.



3.3 Lung function

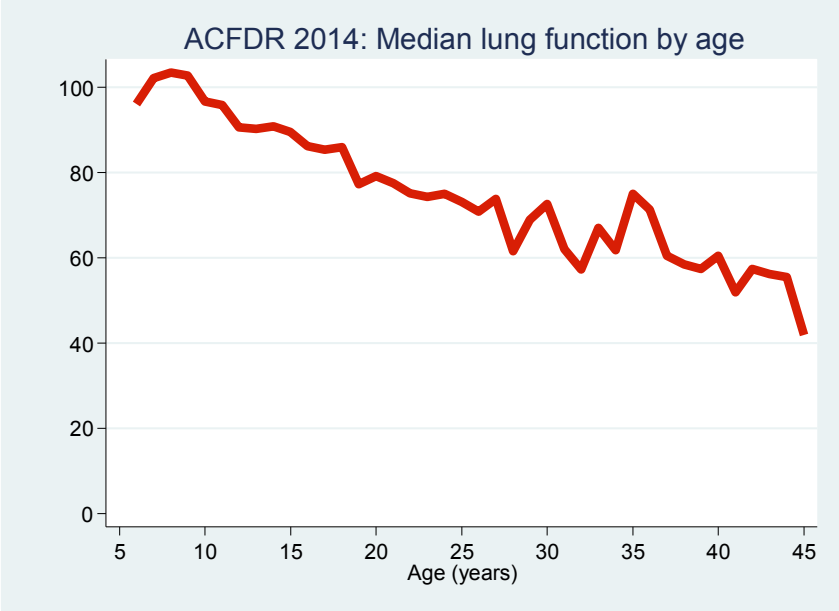
Lung function measures reported here are aligned with methods used in the United States' Cystic Fibrosis Foundation's Patient Registry. That is, the lung function measure included for each patient is the average of the highest FEV1 per cent predicted value recorded in each quarter of the year.

Lung Function categories described in tables and charts:

- Normal – 90% of predicted FEV_1 and above
- Mild impairment – at least 70% but below 90% of predicted FEV_1
- Moderate impairment – at least 40% but below 70% of predicted FEV_1
- Severe impairment – below 40% of predicted FEV_1

Predicted values are based on Global Lung Initiative formulae – see Technical Notes

Median CF lung function, measured as FEV_1 percent predicted, is within the normal range for young children but is lower than 70 per cent of normal, the level at which moderate lung function impairment is experienced, in adult patients aged from around 25 years. Just over 5 per cent of children aged 6 to 11 years have FEV_1 values that are below 70 per cent of predicted values but 13 per cent of older children and adolescents are in this category.

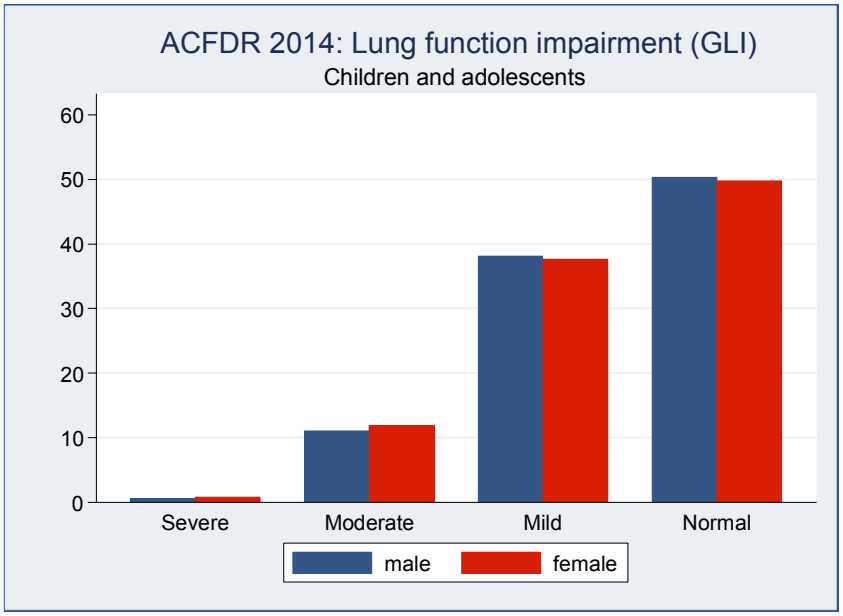


Generally greater proportions of patients have severe lung function impairment in successive older age groups. The proportion of adult male patients with severe lung function impairment (17%) is greater than the proportion of female patients in this severity category (13%). Details for finer age groups in 2014 are in the table on the following page.

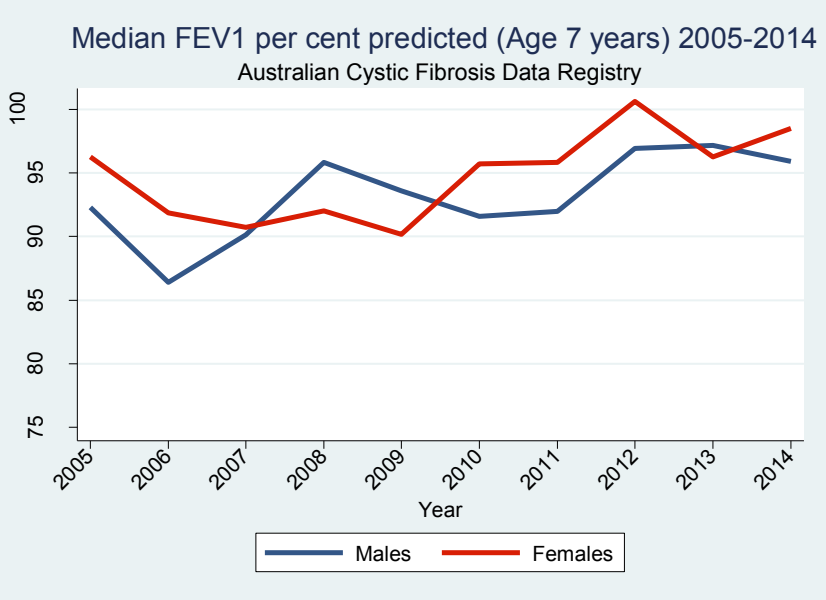
ACFDR 2014: Lung function impairment by age group and sex

	Severe	Moderate	Mild	Normal	Total	Severe	Moderate	Mild	Normal	Total
			Number					*Per cent*		
Males:										
6 - 11 years	1	12	72	170	255	0.4	4.7	28.2	66.7	100.0
12 - 17 years	2	32	100	128	262	0.8	12.2	38.2	48.9	100.0
18 - 29 years	43	147	132	90	412	10.4	35.7	32.0	21.8	100.0
30 + years	88	148	76	29	341	25.8	43.4	22.3	8.5	100.0
Total measured	*134*	*339*	*380*	*417*	*1,270*	*10.6*	*26.7*	*29.9*	*32.8*	*100.0*
Females:										
6 - 11 years	0	15	69	201	285	0.0	5.3	24.2	70.5	100.0
12 - 17 years	4	34	79	111	228	1.8	14.9	34.7	48.7	100.0
18 - 29 years	30	138	106	76	350	8.6	39.4	30.3	21.7	100.0
30 + years	46	118	68	25	257	17.9	45.9	26.5	9.7	100.0
Total measured	*80*	*305*	*322*	*413*	*1,120*	*7.1*	*27.2*	*28.8*	*36.9*	*100.0*
Persons:										
Total measured	*214*	*644*	*702*	*830*	*2,390*	*9.0*	*27.0*	*29.4*	*34.7*	*100.0*

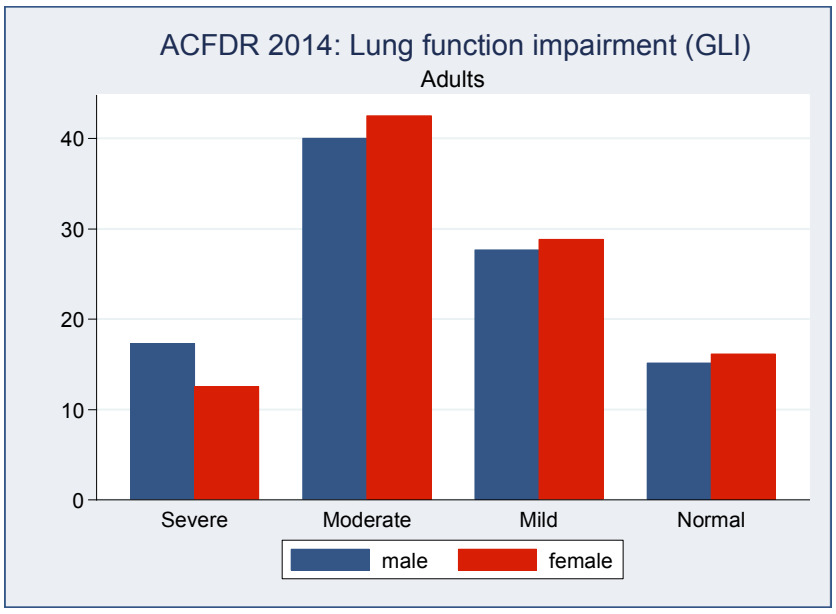
The chart below shows categories of lung function impairment experienced by the child and adolescent CF population as a whole. Fifty eight per cent of male and 61 per cent of female children and adolescents had lung function within the normal range. Consistent with the experience of declining lung function with age, and noting that these are cross-sectional data, the table above shows that proportions of patients with normal lung function are higher for those aged 6 to 11 years than for 12 to 17 year olds, for both males and females.



Upward trends over recent years in child and adolescent age groups were charted in the 2013 Annual Report. The following view, from cross-sectional data for 7 year old children, suggests improvements in early childhood lung care.



For adults with cystic fibrosis, a different pattern of lung function impairment is evident, with just 16 per cent of adult males and 17 per cent of adult females having normal lung function in 2014. Severe lung function impairment was experience by 17 per cent of male adults and 13 per cent of female adults.



Trend data for adult lung function (not shown) indicate improvement for the 18 to 29 years age group since 2005, especially for females. A flatter trend for adults aged 30 and over may be confounded by possible increased survival.

3.4 Nutrition: weight height and body mass index

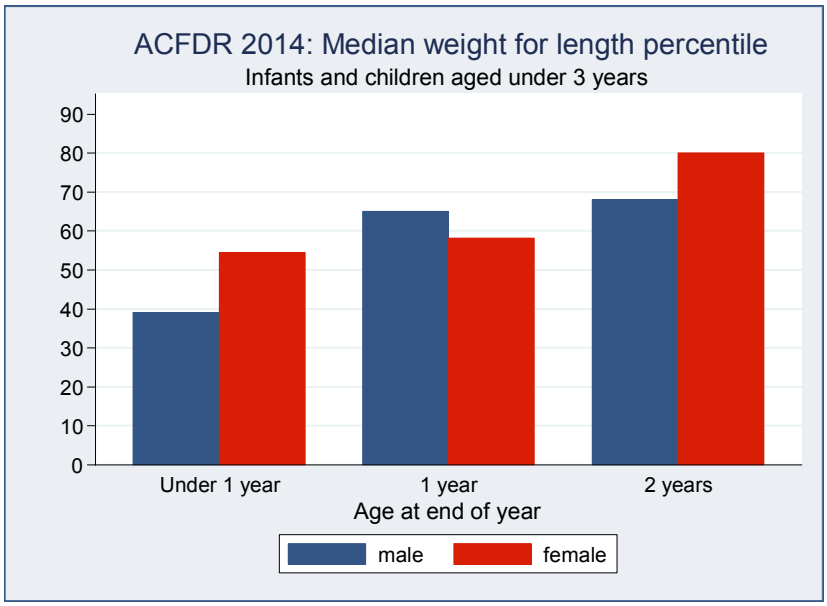
Methodological note

As for lung function measures reported above, values reported in this section are the average of the highest value recorded in each quarter of the year.

Infants and young children aged under 3 years

Nutritional outcomes for children aged under 3 years were introduced for the first time in the 2013 report. For 2014, median value of weight for length is at the 55th percentile for female infants but for males it is at the 39th percentile. This difference may be cohort-specific. Sex differentials at ages 1 and 2 years in 2014 reflect the age pattern of the cohort one year ago, as shown in the 2013 chart.

Percentiles are derived from World Health Organisation Child Growth Standards (WHO 2006) – see Technical Notes.



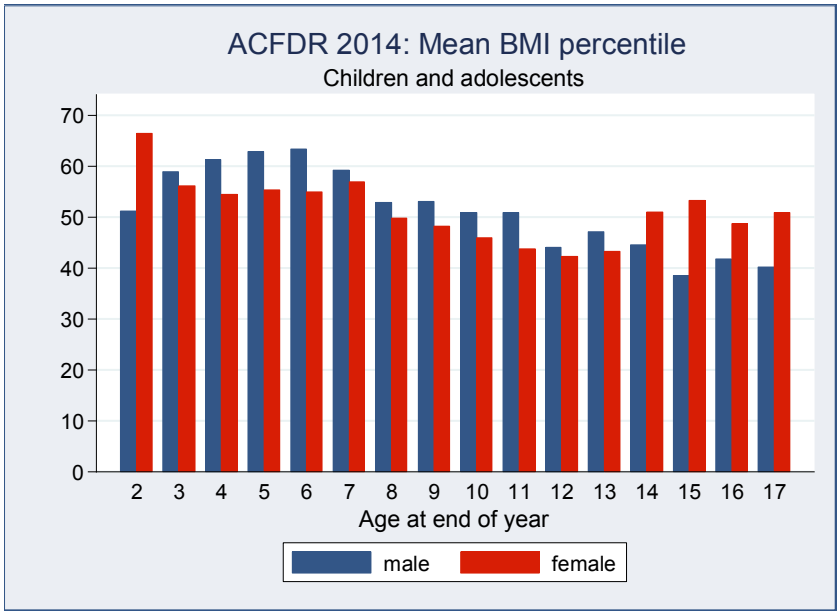
Children and adolescents

Median height percentile for young male children is higher than the reference population, but is below the 50th percentile in older male and all female child and adolescent age groups in 2014. Body mass index (BMI) percentiles are higher than height percentiles for each age and sex group, with the exception of male adolescents, and females in the age group 6 to 11 years, each of whose height and BMI percentiles are at a similar level. The latter group is the only one to not fit a pattern of lower percentiles for both indicators in successively higher age groups.

ACFDR 2014: Child and adolescent height and BMI: median percentiles by age group and sex

	Height	BMI
Males		
2 - 5 years	61.0	63.0
6 - 11 years	42.2	57.4
12 - 17 years	41.3	40.8
Females		
2 - 5 years	45.9	59.5
6 - 11 years	49.6	49.3
12 - 17 years	39.8	48.3

BMI percentiles across individual ages show a generally consistent pattern of lower values at higher ages. For all but the youngest patients (aged 2 years), males under 13 years have higher median BMI percentiles than their female counterparts. Single year age cohorts of adolescent females aged from 14 to 17 years have BMIs around the 50th percentile, higher than their male counterparts.



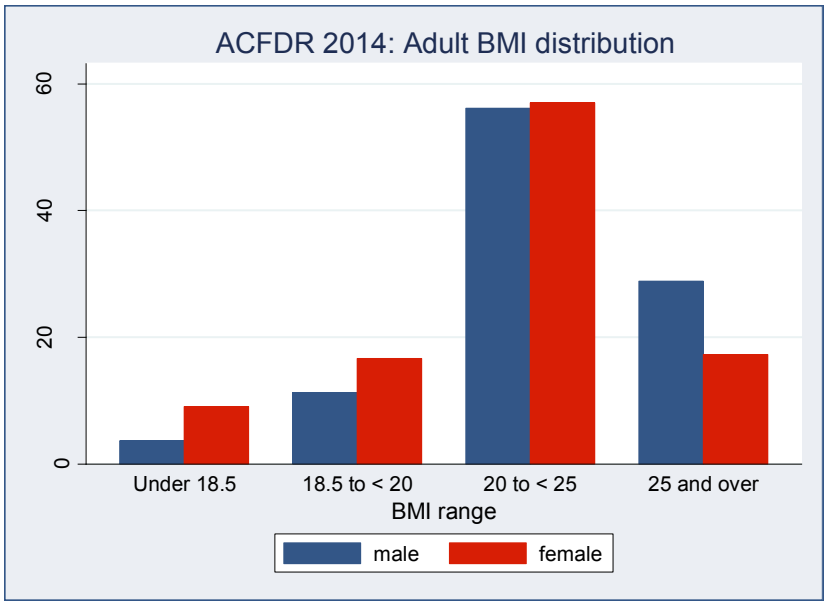
Overall, just under half (47.2 per cent of males and 49.0 per cent of females) were below the 50th percentile for BMI in 2014. The distribution is shown in the table opposite.

ACFDR 2014: Child and adolescent height and BMI percentile distributions by sex

	Height			BMI		
	Males	Females	Persons	Males	Females	Persons
	Per cent			*Per cent*		
< 3rd	3.5	4.8	4.2	2.5	1.2	1.9
3rd - 4.99th	1.8	1.6	1.7	1.3	0.6	1.0
5th - 9.99th	6.0	5.7	5.9	4.0	3.4	3.7
10th - 24.99th	15.3	16.0	15.7	12.4	12.6	12.5
25th - 49.99th	27.1	27.2	27.2	27.0	31.2	29.1
50th - 74.99th	24.7	24.2	24.5	29.5	30.9	30.2
75th - 89.99th	12.2	13.9	13.1	16.8	14.8	15.8
90th - 94.99th	5.0	4.0	4.5	3.5	3.1	3.3
95th - 96.99th	1.8	1.1	1.4	0.9	1.1	1.0
>= 97th	2.5	1.5	2.0	2.2	1.1	1.6
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
	Number			*Number*		
Total	*679*	*669*	*1,348*	*679*	*669*	*1,348*

Adult body mass index

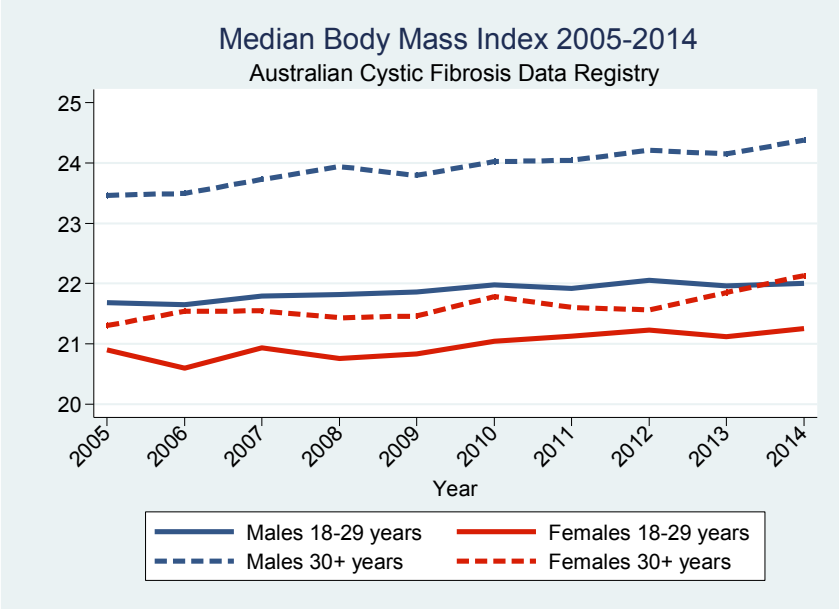
Adult body mass index scores show 56.2 per cent of males and 57.0 percent of females had an average quarterly BMI score in the range 20 to less than 25. The proportion of females who had BMI scores below 20 (25.7 per cent) is higher than the proportion of males (15.0 per cent). Over a quarter (28.9 per cent) of adult males had a BMI above 25.



ACFDR 2014 Adult BMI distribution

	BMI range				
	Less than 18.5	**From 18.5 to <20**	**From 20 to <25**	**25 and over**	**Total**
			Males: per cent		
18 - 29 years	5.1	17.6	60.2	17.1	100.0
30 + years	2.0	4.0	51.4	42.6	100.0
Male adults measured	*3.7*	*11.3*	*56.2*	*28.9*	*100.0*
			Males: number		
Male adults measured	*28*	*86*	*428*	*220*	*762*
			Females: per cent		
18 - 29 years	11.3	17.5	57.1	14.1	100.0
30 + years	6.0	15.5	57.0	21.5	100.0
Female adults measured	*9.1*	*16.6*	*57.0*	*17.3*	*100.0*
			Females: number		
Female adults measured	*56*	*103*	*353*	*107*	*619*

The following chart shows general increases in median BMI values for grouped adult age data since 2005.



4 Treatment of cystic fibrosis

This Chapter describes the treatments and therapies recorded for patients in the Australian Cystic Fibrosis Data Registry.

4.1 Visits to clinics

The average number of clinic visits during 2014 was 4.6 for children and adolescents and 4.9 for adults. These figure should be treated with some caution as they may have been affected by different practices in recording clinic visits at contributing centres. The median number of visits to clinics is less likely to be so affected, and stood at 4 for children and adolescents and also the same for adults in 2014.

4.2 Therapy for cystic fibrosis patients

Antibiotic therapy was prescribed for 94.1 per cent CF patients overall, and for more than 90 per cent of patients in each age group. These proportions, and those that follow in this section, were compiled from therapy usage information supplied for 2,143 patients, 65.1 per cent of all patients in the Registry. Three paediatric centres and four adult centres had missing data in 2014. Proportions of child or adolescent, and adult patients for whom therapy data were missing were 30.1 and 39.5 per cent respectively.

Oral antibiotic therapy was prescribed for 92.6 per cent of antibiotics users. Both PRN (as needed) and continuous use was prescribed for these patients at some time during 2014, as shown in the following table. Higher proportions of adolescents (39.2%) and very young children (45.9%) than those in other age groups were prescribed oral antibiotics for continuous use.

ACFDR 2014: Oral antibiotic therapy - mode of use by age group [(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Mode of use:							
As needed (PRN)	48.0	75.2	74.5	68.8	86.5	87.3	77.7
Continuous	53.0	30.1	30.0	41.1	36.2	34.7	35.7
Mode of use unknown	0.0	0.0	0.0	0.3	0.0	0.6	0.2
Total oral antibiotics users(b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total oral antibiotics users	100	206	377	321	467	346	*1,817*

(a) Patients for whom no treatment information was provided (35 per cent of total) were excluded from analysis.
(b) More than one mode of use can be recorded so numbers add to more than 100.0. As well, mode of use was not recorded for all patients where oral antibiotics were reported.

Just over one half (52.4%) of antibiotics users used inhaled antibiotics in 2014, with proportions generally greater in successively older age groups.

ACFDR 2014: Inhaled antibiotics by age group[(a)]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
				Per cent			
Inhaled antibiotics							
Yes	14.6	25.7	39.4	54.2	66.4	68.7	52.4
No	63.1	54.4	39.7	29.3	22.6	19.2	31.9
Unknown	5.8	2.4	1.3	2.4	0.0	0.8	1.4
Total antibiotics users	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total antibiotics users	*103*	*206*	*388*	*334*	*541*	*390*	*1,962*
Mode of use							
As needed (PRN)	86.7	81.1	75.8	49.2	57.9	61.2	61.5
Continuous	6.7	15.1	24.8	47.5	45.4	42.5	39.8
Mode of use unknown	0.0	1.9	0.0	0.0	0.8	0.7	0.6
Total inhaled antibiotics users(b)	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total inhaled antibiotics users	*15*	*53*	*153*	*181*	*359*	*268*	*1,029*

(a) Patients for whom no treatment information was provided (35 per cent of total) were excluded from analysis.
(b) More than one mode of use can be recorded so numbers add to more than 100.0. As well, mode of use was not recorded for all patients where oral antibiotics were reported.

Almost all CF patients use a range of other therapies to manage conditions other than infections, and many take nutritional supplements. Therapies used by the highest proportion of patients include pancreatic enzymes (81.3% of children and adolescents and 81.9% of adults), vitamin supplements (71.8% and 73.8% respectively), bronchodilators (34.0% and 66.5%) and salt tablets (41.8% and 25.0%). Mannitol was a new addition to the list of therapies reported for 2014.

ACFDR 2014: Other therapy by type[(a)(b)]

	Child/adolescent		Adult	
	Number	Per cent	Number	Per cent
Dornase alpha	468	41.6	525	51.6
Pancreatic enzymes	915	81.3	834	81.9
Vitamin supplements	808	71.8	751	73.8
Bronchodilators	383	34.0	677	66.5
Corticosteroids inhaled	201	17.9	403	39.6
Corticosteroids oral	64	5.7	95	9.3
Mannitol	56	5.0	61	6.0
Insulin	81	7.2	224	22.0
Macrolides	101	9.0	643	63.2
Salt tablets	470	41.8	254	25.0
Antihypercalcaemics	7	0.6	38	3.7
Gastric acid secretion reducers	252	22.4	405	39.8
Other	611	54.3	402	39.5
Patients with therapies reported	*1,125*	*100.0*	*1,018*	*100.0*

(a) Patients for whom no treatment information was provided (35 per cent of total) were excluded from analysis.
(b) Individuals may use more than one type of therapy; percentages by type of therapy add to more than 100.0.

ACFDR 2014: Nutritional supplements by age group[(a)(b)]

	0 to 1 year	2 to 5 years	6 to 11 years	12 to 17 years	18 to 29 years	30+ years	All ages
				Per cent			
Oral (prescribed)	17.8	23.9	27.8	23.9	29.3	29.6	27.3
Nasogastgric	6.8	2.6	1.7	2.5	2.0	2.4	2.4
Total Parenteral Nutrition (TPN)	2.7	0.0	0.0	0.0	0.2	0.0	0.2
Gastrostomy tube/button	0.0	3.2	6.6	12.9	4.5	1.7	5.3
Nutritional supp. type unknown	0.0	0.0	0.0	0.0	0.0	0.0	0.0
Total using nutritional supplements	21.9	27.7	33.8	35.7	33.3	31.1	32.3
Not using nutritional supplements	78.1	72.3	66.2	64.3	66.7	68.9	67.7
Patients with nutritional supplements reported	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
				Number			
Total with nutritional supplements. reported	*73*	*155*	*302*	*280*	*552*	*409*	*1,771*
Nutritional supps. not reported	42	67	118	88	38	19	372
Patients with therapies reported	*115*	*222*	*420*	*368*	*590*	*428*	*2,143*

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.
(b) Individuals may use more than one type.

Of the 2,143 patients for whom treatment data were reported in 2014, 20 were reported to have commenced oxygen therapy during 2014 and 19 remained on oxygen therapy commenced in a previous year. The majority in each category (19 (95%) and 16 (84%) respectively) were adults.

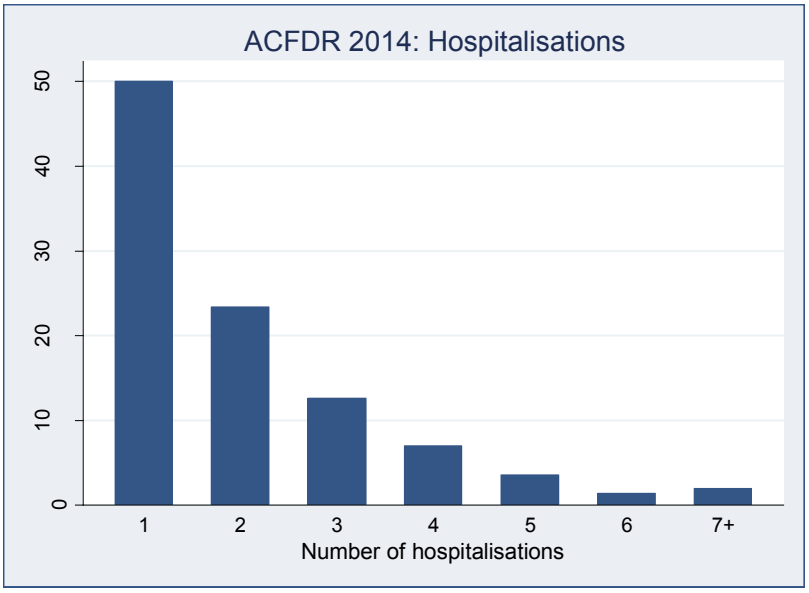
Twelve patients commenced using non-invasive ventilation in 2014 and 20 had commenced in earlier years. All but one of those commencing during 2014 and two who had commenced earlier were adults.

For both non-invasive ventilation and oxygen therapy, it is likely that the numbers reported are incomplete, as the full patient population of two adult CF centres is missing from reported data.

4.3 Hospital treatment

The manner of collection of hospitalisation data for the Registry does not allow a clear distinction to be drawn between patients with 'no hospitalisation' and those who have missing data. Four paediatric hospitals (including 2 large, one medium size and one small in patient numbers) and four adult hospitals (three large and one medium) were excluded from the analysis of hospitalisation because no data or incomplete data were provided. This excluded 39 per cent of patients in the Registry from this analysis.

Of the 2,012 patients attending hospitals that provided adequate data, 45.8 per cent experienced at least one hospitalisation for any indication during 2014. Half of these had more than one period in hospital during the year, as the chart below shows.

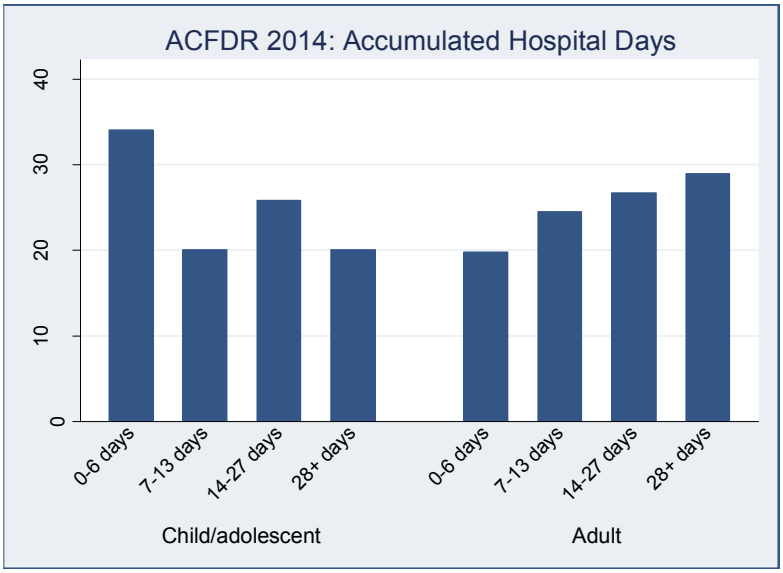


ACFDR 2014: Hospitalisation related to cystic fibrosis, respiratory causes[a]

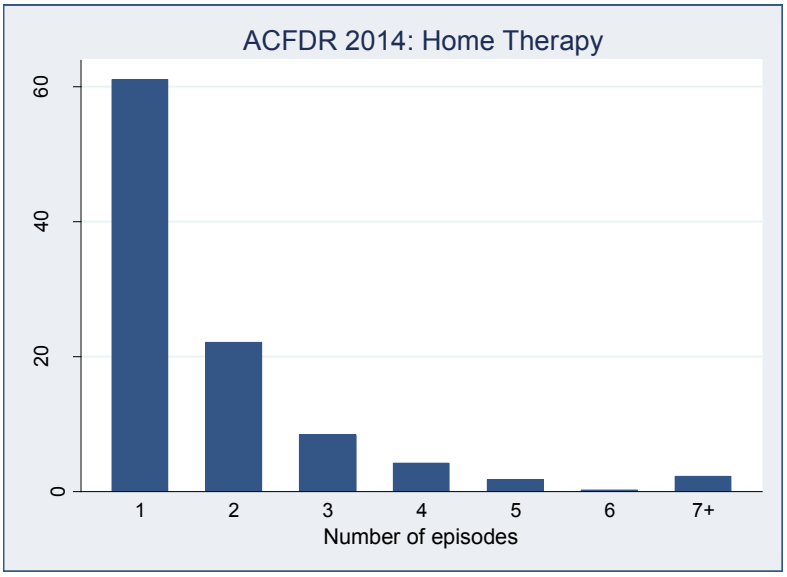
	Persons aged						
	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	All ages
	Per cent of persons in age group						
Number of hospitalisations:							
None or none reported	46.9	51.7	61.8	48.6	49.8	60.3	54.2
1	24.0	33.0	20.7	25.1	25.1	21.1	24.2
2	15.6	10.3	9.2	13.4	11.1	9.3	10.9
3	8.3	2.5	3.9	5.7	6.4	3.7	4.9
4	1.0	2.0	2.4	3.1	3.6	3.3	2.9
5	3.1	0.0	0.8	2.3	1.5	1.9	1.5
6	1.0	0.5	0.3	0.6	1.5	0.2	0.7
More than 6	0.0	0.0	1.1	1.1	1.1	0.2	0.8
Total	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*	*100.0*
	Number of persons						
Total	96	203	382	350	550	431	2,012

(a) Eight CF centres were excluded from analysis (see text)

Of the 1,013 persons who were hospitalised in 2014, just over half (50.6%) accumulated at least 14 admitted days through the year. Mean and median days, of 20 and 14 respectively for these people, underline the fact that some CF patients spend considerable periods of time hospitalised. Adult patients (mean 23.8 days, median 14 days) generally spent more days as admitted patients in hospital than children adolescents (16.5 and 12 days respectively)



4.4 Home therapy

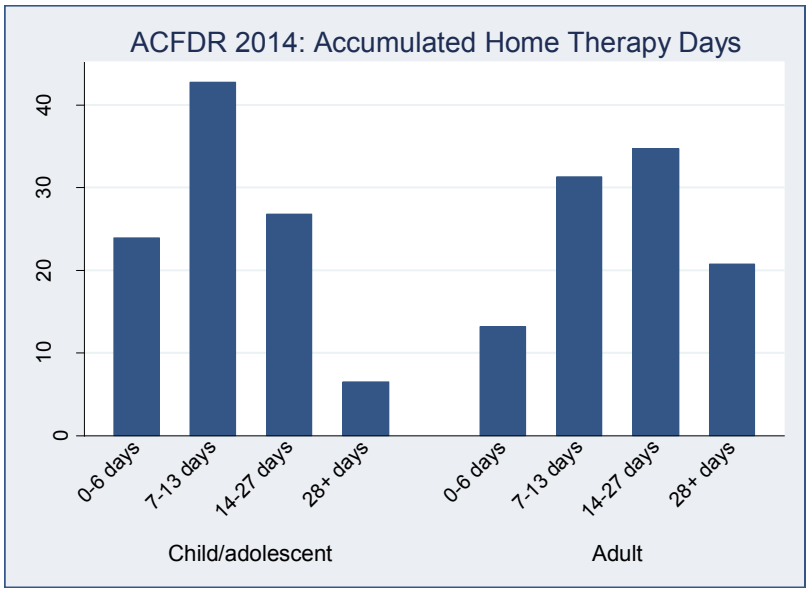


As for the hospitalisation data, the manner of collection of data about intravenous antibiotic therapy administered at home does not allow a clear distinction to be drawn between 'no home therapy' and missing data in relation to a patient. Five paediatric hospitals (including 2 large, one medium size and two small in patient

numbers) and four adult hospitals (two large and one medium and one small) were excluded from the analysis of home therapy because no data or incomplete data were provided. This excluded 36 per cent of patients from the analysis

A total of 133 child or adolescent patients and 261 adult patients in the population analysed spent a mean of 13.8 days and 20.3 days respectively under intravenous antibiotic therapy at home. Median days were 10 and 14 respectively. The following chart shows the distributions of those days.

The mean numbers of episodes for persons undergoing home therapy were 2.7 for children or adolescents and 5.7 for adults. Episodes of home therapy may be preceded by a short period of hospitalisation



4.5 Non-transplant surgery

The following table shows the age distribution of persons reported as having undergone selected non-transplant surgery during 2014. In view of the incompleteness of reporting, these numbers are likely to be under-estimates.

ACFDR 2014: Non-transplant surgery during the year[a]

	0 - 1 years	2 - 5 years	6 - 11 years	12 - 17 years	18 - 29 years	30 + years	Total
IV access devices	0	1	9	15	10	9	44
Gall bladder disease	0	0	0	1	5	1	7
Gastrostomy	0	1	8	4	2	2	17
Intestinal obstruction	2	0	1	2	2	1	8
Nasal (any surgery)	0	1	3	3	5	4	16
Other	7	13	13	18	34	29	114

(a) Patients for whom no treatment information was provided (29 per cent of total) were excluded from analysis.

5 Organ transplants

5.1 Patients assessed for transplant in 2014

Cystic fibrosis centres reported 22 patients had been assessed for organ transplant during 2014. Of these, 13 (12 adult) had been accepted onto transplant waiting lists.

Nine patients had been accepted for a bilateral lung transplant and one for a liver transplant. For three other patients accepted, the organ to be transplanted was not specified. Of the nine accepted for lung transplant, seven were reported as having received a transplant during 2014 and are included in the table below.

5.2 Transplants during 2014

Thirty four bilateral lung transplants were reported by CF centres as having occurred in 2014. The majority of these transplants (21) were performed on patients aged 30 years and over.

ACFDR 2014: Patients receiving lung transplants in 2014

Age group:	Males	Females	Persons
12 - 17 years	0	1	1
18 - 29 years	6	6	12
30 years and over	11	10	21
All ages	*17*	*17*	*34*

One female patient was reported as receiving a liver transplant in 2014.

6 Mortality

6.1 Deaths recorded in 2014

There were 19 deaths reported to the Registry in 2014, half the number reported in 2013 when the number of deaths was higher than had been reported in any year since 2000. Two of the deaths reported in 2014 were of persons aged less than 18 years, one male and one female patient of adolescent age.

ACFDR 2014: Deaths, by age and sex

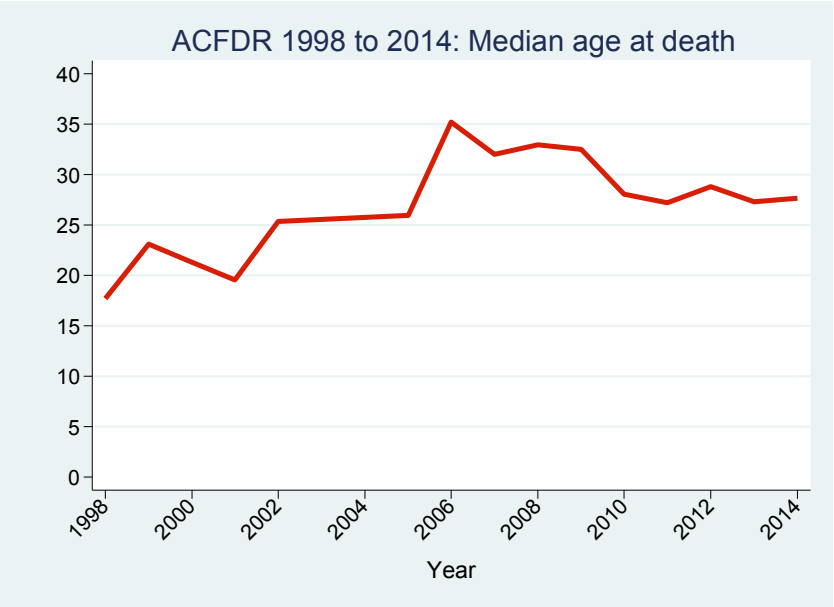
Age group:	Males	Females	Persons
12 - 17 years	1	1	2
18 - 29 years	5	3	8
30 + years	4	5	9
All ages	*10*	*9*	*19*

By state and territory of residence, the highest number of deaths was reported for people residing in New South Wales (7). Five deaths were reported in Victoria, three in Queensland and two in each of South Australia and Tasmania. No deaths of persons with CF in Western Australia were reported in 2014.

ACFDR 2014: Deaths recorded by state

	Males	Females	Persons
New South Wales	4	3	7
Victoria	1	4	5
Queensland	2	1	3
South Australia	2	0	2
Tasmania	1	1	2
Total	10	9	19

The median age at death for patients who died on 2014 was 27.7 years. A rising trend shown in this indicator from 1998 appears to have stabilised or even reversed from around 2006, when the median age at death was 35 years. Such apparent trends should be interpreted conservatively, because of the relatively small number of deaths each year.



6.2 Causes of death

Eleven of the 19 deaths reported in 2014 were due to pulmonary causes.

ACFDR 2014: Cause of death

	Males	Females	Persons
Related to CF:			
Pulmonary	8	3	11
Other (including post-transplant)	1	4	5
Unrelated to CF	0	1	1
Cause unknown or not stated	1	1	2
All causes	*10*	*9*	*19*

Notes

Supplementary tables and technical notes

A range of supplementary tables and technical notes may be accessed on the Cystic Fibrosis in Australia website: http://www.cysticfibrosis.org.au/data-registry.

Technical notes cover:

- Data collection and editing
- Collection instrument
- Identification and resolution of duplicate records
- Derivations for age, lung function and nutrition data
- Registry data quality

Detailed information about data elements, coding schemes and methodology can be provided on request to CFA.

Access to Registry data

Requests for additional information from the Australian Cystic Fibrosis Data Registry are welcome. Application should be made to Cystic Fibrosis Australia (CFA). In accordance with a CFA policy on charging for ACFDR data services, a fee may be charged to recover costs.

Researchers proposing to undertake analysis of unit records may be granted access to de-identified patient records, subject to approval by the Registry's medical advisory and ethics committees, and to researcher agreement to CFA's conditions of use. Interested researchers are advised to contact CFA for details and to arrange consideration of their research proposal.

All communication about additional data requirements and research access should be addressed to:

Nettie Burke
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PO Box 268
North Ryde NSW 1670

Phone: +61 (0)2 9889 5171
Email: nettieb@cfa.org.au

Abbreviations

ACFDR	Australian Cystic Fibrosis Data Registry
BAL	Bronchi alveolar lavage
BMI	Body mass index
CF	Cystic fibrosis
CFA	Cystic Fibrosis Australia
FEV1	Forced expiratory volume (litres) in 1 second
GLI	Global Lung Initiative
MRSA	Methicillin-resistant *Staphylococcus aureus*
TPN	Total parenteral nutrition